

Nutritional management of an adult with Phenylketonuria with the use of a Glycomacropeptide-based PKU formula

Glycomacropeptide (GMP) is a dietary peptide which is inherently low in Phenylalanine. GMP-based formulas offer a promising alternative to traditional amino acid (AA) based formulas for the dietary management of Phenylketonuria (PKU). Some individuals with PKU prefer GMP-based formulas as they feel the products have a better taste over AA-based. They also enjoy that GMP-based formulas contain a source of more intact protein and may possibly provide a feeling of fullness for longer. This case report reviews the successful introduction of a GMP-based formula (*PhenylAde® GMP Drink Mix*) in an adult female with classical PKU.

History

This patient is a 40-year old female diagnosed with classical PKU shortly after birth. She has been on a low protein diet since diagnosis and has been mostly compliant throughout life with both her diet and PKU formula; except for a slight relaxation in diet during her teenage years. She adhered flawlessly to a preconception and pregnancy diet during her mid-20s and has since continued to follow the PKU diet with regular contact with her metabolic clinic. She has a target blood phenylalanine (PHE) range of 120-700 $\mu\text{mol/L}$ (2-11.7 mg/dL), and her blood spot results are typically within range at $<500 \mu\text{mol/L}$ ($<8.3 \text{ mg/dL}$). She has a secondary diagnosis of hypothyroidism for which she is prescribed thyroid hormone medication. She also experiences daily gastrointestinal reflux for which she has been prescribed an antacid.

She is a healthy weight (59.5 kg) with a BMI of 21.3 kg/m^2 . Her estimated total protein requirement is 60 g/d and advised to consume no more than 300 mg of PHE per day.



“Very easy- although quite a lot to drink - the taste is so much more pleasant”

Nutritional Management

She has been on a standard AA-based formula for several years. She receives 60 g protein equivalent (PE) per day, and reports 100% compliance; although reflux is an ongoing issue. To start a GMP-based formula the patient was prescribed 30 g PE/d of *PhenylAde® GMP Drink Mix* (3 x 33.3 g sachets per day; 10 g PE per sachet), which provided 50% of her total protein requirements and replaced one of her previously prescribed AA-based formulas. As *PhenylAde® GMP Drink Mix* contains some residual PHE (15.3 mg PHE/10 g PE), she was advised to reduce her PHE intake from food by an amount roughly equivalent to the PHE provided in 3 sachets (~45 mg PHE). *

*Practices vary from clinic to clinic.

Results

Over a 4-week period of taking *PhenylAde® GMP Drink Mix* the patient had 100% compliance and tolerated the GMP-based formula perfectly, with no adverse events. She reported less reflux and coughing. She also reported reduced feelings of hunger which she said resulted in less sugar cravings and snacking. She likes the taste of *PhenylAde® GMP Drink Mix* and found it very easy to drink. She prefers *PhenylAde® GMP Drink Mix* in terms of both taste and tolerance over her usual AA-based formula. Comments from this individual included:

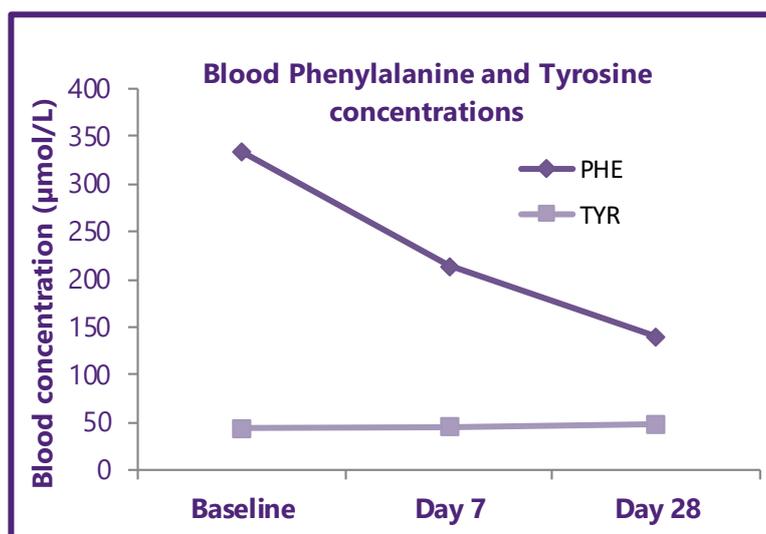
“Now that I’ve got into a routine, I’m finding it very easy to make and drink”

“Very easy- although quite a lot to drink - the taste is so much more pleasant”

“It’s so good not to have permanent bad breath/repeating amino acids and I’m coughing less”

Blood amino acid concentrations

Her blood PHE levels decreased from 314 $\mu\text{mol/L}$ (5.2 mg/dL) (Baseline) to 214 $\mu\text{mol/L}$ (3.6 mg/dL) (Day 7) and then to 140 $\mu\text{mol/L}$ (2.3 mg/dL) (Day 28); though this may have been due to a slight decrease in overall protein intake from food. Her blood tyrosine (TYR) concentration remained stable from 44 $\mu\text{mol/L}$ (0.73 mg/dL) (Baseline) to 45 $\mu\text{mol/L}$ (0.75 mg/dL) (Day 7) and 48 $\mu\text{mol/L}$ (0.8 mg/dL) (Day 28). Her branched-chain amino acid (BCAA) concentrations also remained within population reference ranges throughout the 4-weeks.



Conclusion

This female successfully introduced *PhenylAde*[®] *GMP Drink Mix* into her diet for 28 days with no adverse outcomes. She reported a greater preference for the GMP-based formula and better tolerance compared to her usual AA-based formula. Her blood AA levels remained within safe ranges and the Metabolic Dietitian managing her reported that all goals she had set had been met. The patient requested to continue *PhenylAde*[®] *GMP Drink Mix* after the 4-week period and currently uses it as half of her daily PE requirement.

The data for this case report was provided by a UK clinician for Nutricia UK